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LETTER

Unexpected hemophagocytic syndrome in a post-cardiac surgery patient

Lisa Linthorst^{*1}, Heleen Aardema² and Jaap E Tulleken²

See related research by Beutel et al., <http://ccforum.com/content/15/2/R80>

In a recent issue of *Critical Care*, we read with interest the article on the incidence of hemophagocytic lymphohistiocytosis (HLH) in patients with H1N1 infection [1]. Acquired HLH is associated with infections (particularly viral), malignancy, and auto-immune disorders [2].

We describe a case of HLH in a 66-year-old woman admitted to the intensive care unit after aortic valve replacement. She had a history of a cerebral vascular event and diabetes mellitus type 2. After surgery, she had an unexplained increase in oxygen demand and a fever. Standard antibiotics were started for probable ventilator-associated pneumonia and were discontinued after negative cultures. A cardiac evaluation showed good valve patency.

Laboratory tests showed elevated creatine kinase (3,562 U/L), lactate dehydrogenase (1,380 U/L), ferritin (3,960 µg/L), and triglyceride (2.29 mmol/L) and decreased hemoglobin (64 g/L), platelets (54×10^9 /L), fibrinogen (1.0 g/L), and haptoglobin (<0.2 g/L). The results of a Coombs test were negative. Schistocytes were absent. Bone marrow aspiration showed marked hemophagocytosis. An eliciting factor could not be found. Because criteria for HLH were met, we started prednisone treatment. Our patient made a satisfactory recovery.

Fever, thrombocytopenia, and anemia are common after cardiac surgery. Without the finding of a markedly elevated ferritin level in our patient, the proper diagnosis could have been missed or her condition could have been misclassified as sepsis with multiple organ failure [3]. To the best of our knowledge, this is the first published case of HLH to occur after cardiac surgery but without a known eliciting cause. This raises a tantalizing thought:

unknown triggers may have a role in the pathogenesis of HLH. It would be interesting to search for HLH in a cohort of cardiac surgery patients.

Abbreviation

HLH, hemophagocytic lymphohistiocytosis.

Competing interests

The authors declare that they have no competing interests.

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Our patient provided written consent to the publication of this letter.

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